

Giant Pancreatic Retention Cyst Secondary to Chronic Pancreatitis: Report of a Case

D ZOSIMAS, PM LYKLOUDIS, R KUMAR, A SHANKAR

Abstract

Purpose: To report a rare case of massive pancreatic duct dilatation. **Methods:** Case report of a 16-year-old female with complex medical background presented acutely in our institution after complicated endoscopic drainage of a symptomatic giant (>16 cm) cyst of the head of the pancreas. **Findings:** Following a laparotomy the cyst was drained by means of a Roux-en-Y cysto-jejunostomy. Histopathology revealed a true pancreatic cyst. **Conclusions:** Retention cysts usually are uncomplicated and differentiation from congenital cysts is difficult. In this case, presence of pancreatic calculi, age of diagnosis and recurrent episodes of acute pancreatitis were in favour of a retention pancreatic cyst. A cyst of this size and association of true pancreatic cysts with chronic pancreatitis and pancreatic calculi has never been reported previously.

Key words

Chronic pancreatitis; Cystojejunostomy; Retention cyst; True pancreatic cyst

Introduction

Pancreatic cystic lesions are uncommon but not rare. Pseudocysts comprise up to 70% of such lesions. True (simple) retention pancreatic cysts are less frequently seen and can be associated with chronic pancreatitis, possibly related to an increased incidence of stone formation.¹

The authors present an interesting and unique case of chronic pancreatitis associated with a massively (>16 cm) dilated pancreatic duct-retention cyst in a 16-year-old female.

Methods

The patient was admitted in our institution with peritonitis two days after an endoscopic cysto-gastrostomy and stent insertion for large pancreatic cyst. Pre-endoscopy Magnetic Resonance Imaging demonstrated the lesion to measure 20x11 cm. She had background of end-stage renal failure secondary to Leber's congenital amaurosis associated with nephropathy (Senior-Löken syndrome),² treated by a renal transplant in 2006. This was complicated by delayed graft function, acute tubular necrosis and a subsequent laparotomy with wash out. In 2004 the patient had a percutaneous endoscopic gastrostomy inserted, which was exchanged in 2007 and 2012. In 2005 she underwent a laparoscopic cholecystectomy and in 2006 an abdominal hernia repair. There was also background of Caroli's disease and presence of multiple intrahepatic calculi and strictures as well as pancreatic atrophy, associated with dilated pancreatic duct, calcifications, and obstructing calculi of the head of pancreas. At least seven episodes of acute pancreatitis were documented, starting from 2000, with diagnosis of dilated pancreatic duct-retention cyst made in 2002. Surveillance

HPB Surgery & Liver Transplantation, Royal Free Hospital, London, UK

D ZOSIMAS	MD
PM LYKLOUDIS	MD, MSc, DIC
R KUMAR	FRCS
A SHANKAR	FRCS

Correspondence to: Dr D ZOSIMAS

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imaging demonstrated increasing size of the cystic lesion. An episode of cardiac arrest was also documented during an intensive care unit admission. At the time the patient arrived, clinical examination revealed abdominal distension and diffuse abdominal tenderness. A computerised tomography (CT) scan revealed a large amount of free intraperitoneal air and fluid and also the distal end of the stent lying just superior to the tail of pancreas, having been displaced. The decision was made to proceed with urgent laparotomy through a midline incision. Pus was found in the lesser sac together with two slipped endoscopic stents. Washout was performed, the stents were removed and their exit site on the posterior gastric wall was closed. The massively dilated pancreatic duct was de-roofed and tissue sent for histology (Figure 1). A definitive drainage procedure was performed by way of a cysto-jejunostomy, utilising a retrocolic Roux loop of jejunum, and a jejuno-jejunostomy.

Results

The immediate recovery was uncomplicated, other than pain management. The microscopic examination of the specimen revealed a cyst wall composed of fibrovascular and fatty tissue with vascular congestion. In many areas the cyst was lined by a single layer of epithelium with abundant eosinophilic cytoplasm and round, bland nuclei. In places residual pancreatic tissue was noted, comprising ductular structures lined by bland epithelium and pancreatic acini. There was no evidence of dysplasia or malignancy. The conclusion was of a true non-neoplastic pancreatic cyst.

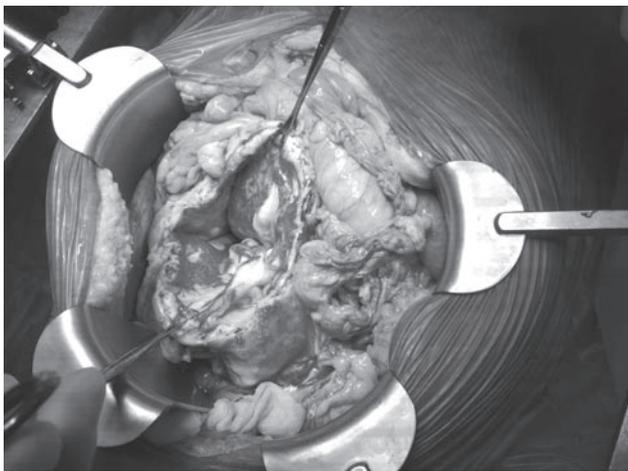


Figure 1 Intraoperative image depicting the large pancreatic cyst opened.

During the next three weeks the patient was complaining of vague abdominal symptoms and developed diarrhoea and anaemia. In consecutive imaging scans the pancreatic duct was markedly decreased in size in the body and tail of pancreas but unchanged in the head and no drainable abdominal collections were demonstrated. The intrahepatic biliary tree was abnormal and with bilateral cholangiopathy present which could partially justify the clinical picture. A further CT scan (Figure 2) in the fourth post-operative week revealed increased cystic dilatation of the pancreatic duct, stable appearances of the bile ducts and colitis. A subsequent culture demonstrated *Clostridium Difficile* colitis which was treated successfully with oral antibiotics and stool cultures on discharge were negative. Based on the complexity of the case and the psychosomatic peculiarities of the patient, after an intraabdominal leak was excluded, she was transferred to a specialised paediatric unit for further care. Treatment continued conservatively without further complications. A subcutaneous wound infection healed well. Two subsequent ultrasound scans showed the cyst decreasing in size and she was discharged 2 weeks later. The exocrine enzymatic function tested on discharge was: Amylase: 138 U/L, Lipase: 245 U/L. In the latest follow up after 12 months the patient was well, although experiencing abdominal pain recurrences related to chronic pancreatitis. Her feeding is being absorbed



Figure 2 Enhanced computerised tomography section depicting dilated pancreatic duct in the area of head of pancreas (black arrow), the non-distended pancreatic duct in the area of the body and tail of pancreas (black arrowhead) and the jejunal loop used for cysto-jejunostomy (white arrowheads).

sufficiently and no signs of established pancreatic insufficiency exist.

Discussion

Pancreatic pseudocysts are the most common non-neoplastic cysts of the pancreas.³ Other non-neoplastic, benign cysts are rare and include retention cysts, congenital-developmental cysts, duplication cysts, mucinous non-epithelial cysts and parasitic cysts. The first three types are also known as true cysts and have a true epithelial component to their wall.⁴

Retention cysts are gross enlargements of the pancreatic duct and contain a lining of endothelial cells, distinguishing them from pseudocysts whose wall consists of dense fibrous tissue.¹ Responsible for their formation is the obstruction of a normal duct by different causes like stones, inflammation, trauma, fibrous strictures, mucus plugs in cystic fibrosis (pancreatic cystosis) or tumours and neoplastic epithelial proliferations^{1,3} (retention cysts have been reported in up to 15% of patients with pancreatic carcinoma).⁵

Complications of retention cysts are unexpected.^{1,3} Perforation of a retention cyst has only been reported as an iatrogenic complication.⁶ The decision to intervene on the basis of the symptoms and the condition of the cyst has always been controversial.⁷ Therefore, surgical excision is not often required, unless there are persistent symptoms or difficulties in distinguishing from pancreatic malignancy.³ Many centres suggest surgical resection for patients who are fit for surgery and have cystic lesions of the pancreas, given the diagnostic difficulties encountered.³ In our case, the general medical background and the complex medical history of the patient initially imposed a conservative approach, but the persistent chronic abdominal pain and nausea and the recurrent bouts of acute pancreatitis led to endoscopic internal drainage.

Generally, it is difficult to differentiate retention cysts from congenital true cysts by histopathological features.⁴ Usually a retention cyst can be diagnosed preoperatively if it appears as a small cystic lesion communicating with the pancreatic duct and if also a cause of proximal duct obstruction can be seen.³ In our case, pancreatic stones caused the proximal duct obstruction. We believe that the intrahepatic stones have been the cause of recurrent acute pancreatitis which has finally become chronic with calcific obstructions of the head of the pancreas. Primary pancreatic duct pathology was previously excluded. The common bile duct had been normal in diameter in most of recent scans

although in the past was dilated in the distal part during the acute attacks. Symptoms of congenital cysts are seen generally in patients under the age of two years. Only a few cases have been reported in the literature and most of them were diagnosed before the age of two years.⁴ In our case the cyst was diagnosed at the age of six, which is in favour of a retention cyst. Another way of differentiating the two entities is the cystic fluid enzymatic activity, which is lower in congenital cysts.⁴ In this case the fluid enzymatic activity was expected to be higher than normal but not tested. Furthermore, the history of previous episodes of acute pancreatitis is also in favour of the diagnosis of a retention cyst.⁶

A pancreatic duct with massive dilatation of this size (>16 cm), and in association with chronic pancreatitis and pancreatic stones has never been previously reported. Two other cases of giant pancreatic duct dilatation have been described but one was caused by a minor papilla obstruction in a case of pancreas divisum (15x5 cm),⁸ and the other by an islet cell adenoma obstructing the main duct.⁹

Declaration of Conflict of Interest

There are no declarations of conflict of interest by the authors.

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